ELSEVIER

A 40-YEAR-OLD WOMAN WITH BACK PAIN

BRIAN J. PENROD, MD, NOGAH HARAMATI, MD,
DAVID FRAGER, MD, AND HOWARD D. DORFMAN, MD

CLINICAL DATA

The patient was a 40-year-old woman who presented with intermittent, dull, nonradiating sternal and back pain of 9 months' duration. The pain was provoked and aggravated by motion and change of posture, and worsened when the patient was lying down at night. It steadily increased in severity over time.

IMAGING FINDINGS

Roentgenograms from the time of onset of symptoms revealed a destructive, lytic lesion involving the right pedicle of T3. Radionuclide bone scan at that time revealed increased activity at the level of T3 on the right side. A follow-up examination 10 months later demonstrated destruction of the right pedicle of T3 by a mass (Figure 1). CT scan at the level of T3 (Figure 2) revealed a well-circumscribed mass with a narrow zone of transition destroying the right pedicle.

A right T3 costotransversectomy was performed and the lesion was excised.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis included fibrous dysplasia, osteoid osteoma, aneurysmal bone cyst, plasmacytoma, and eosinophilic granuloma.

PATHOLOGY

Histological examination of the lesional tissue (Figure 3) demonstrated proliferation of spindle cells with bland elongated nuclei within a fibrous stroma. Irregularly shaped bony trabeculae were present, scattered within the stroma. No peripheral osteoblasts were seen surrounding these bony spicules which were composed of woven bone. The appearance of the fibrous stroma and the woven bone was diagnostic of fibrous dysplasia. The single focus of involvement characterized this lesion as monostotic fibrous dysplasia (MFD).

DISCUSSION

Fibrous dysplasia is a fairly common skeletal abnormality in which normal bone is replaced by fibroosseous tissue (1). Not a true neoplasm, it is a dysplastic condition of bone forming mesenchyme in which osteoblasts fail to undergo normal maturation (2). As such, the process is usually self-limited, especially in the monostotic form, and often becomes quiescent as patients pass through puberty (3). Patients with the polyostotic form may exhibit a more progressive course of the disease, with extensive skeletal involvement and deformity, and malignant transformation is recognized (4).

Patients presenting with fibrous dysplasia usually do so within the first three decades of life, many presenting in childhood and adolescence (1, 3, 5–7). The polyostotic form tends to present earlier than the monostotic form. Kransdorf et al. (2) reported a mean age of presentation of 8 years for polyostotic fibrous dysplasia and a 2:1 male predominance in the monostotic form.

MFD is much more common than the polyostotic variety, representing 70 to 85% of reported cases (1–3, 5). The majority of the literature, however, reports on polyostotic disease. This may be due to the greater severity of manifestations and progressive nature of this

From the Department of Radiology (B.J.P., N.H.) and the Section of Orthopedic Pathology (H.D.D.) of the Albert Einstein College of Medicine and Montefiore Medical Center, Bronx, and the Department of Radiology, St. Luke's Roosevelt Hospital (D.F.), New York, New York.

Address reprint requests to: Nogah Haramati, MD, Department of Radiology, Montefiore Medical Center, 111 East 210th Street, Bronx, NY 10467.

Received March 12, 1994; accepted October 20, 1994.

655 Avenue of the Americas, New York, NY 10010

0899-7071/96/$15.00
SSDI 0899-7071(95)00049-V

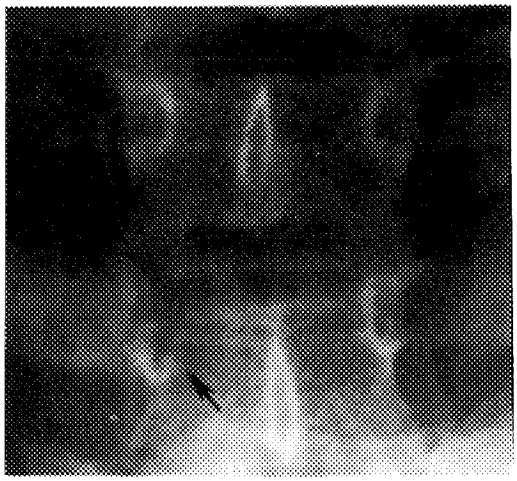

FIGURE 1. Plain film of the lower cervical and upper thoracic regions of the spine demonstrates a lytic lesion destroying the right pedicle of T3 *(arrow)*.

process. Monostotic lesions are not associated with pigmentation and endocrine abnormalities such as the McCune-Albright syndrome (2, 3, 6–9). Malignant transformation is quite uncommon in the monostotic form of the disease (2–4, 6).

The most commonly cited physical deformities are leg length discrepancy and the so-called shepherd's crook deformity of the femoral shaft. However, severe deformity is not typical of the monostotic variety unless the lesion involves a weight-bearing bone (1–3, 6). The entity may present with pain in an extremity, limp, or pathological fracture, or may be completely asymptomatic until it is incidentally discovered (2, 3, 5, 6). Typically, MFD involves the ribs, femur, tibia, craniofacial bones, and humerus, in descending order of frequency (1). The Armed Forces Institute of Pathology (AFIP) reports a small but significant incidence of iliac and radial involvement (2).

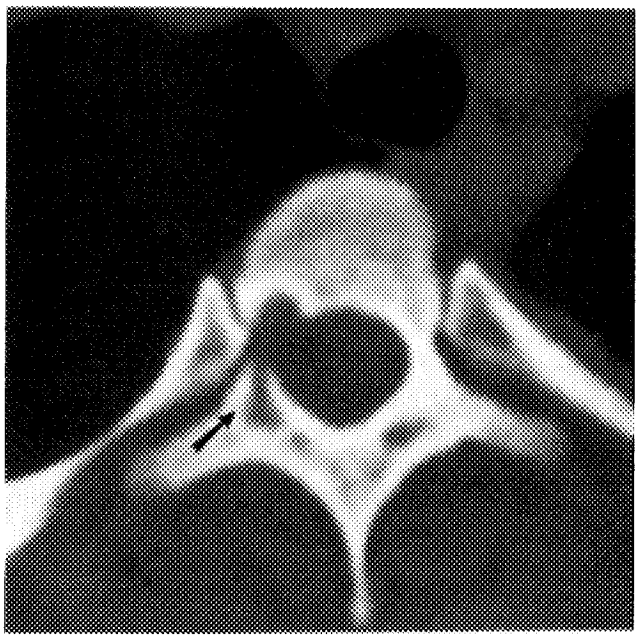

FIGURE 2. Axial CT section through the T3 pedicles reveals a well-circumscribed mass destroying the right pedicle *(arrow)* with a narrow zone of transition, sclerotic rim, and a homogeneous matrix denser than adjacent soft tissue and less dense than bone with no discrete calcifications.

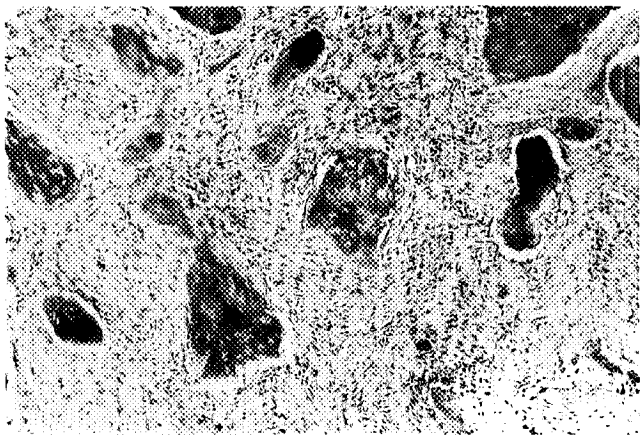

FIGURE 3. Photomicrograph of biopsy specimen reveals a spindle cell proliferation with bland elongated nuclei. Irregularly shaped bony trabeculae are scattered within the fibrous stroma. No peripheral osteoblasts are seen surrounding these bony spicules (H&E stain, original magnification ×100).

Vertebral involvement in MFD is exceedingly rare. Although Mirra et al. (5) reported a 4% incidence of vertebral involvement, cases were not identified. Since Resnik and Lininger (10) reported the sixth pathology-proved case of vertebral MFD in 1984, a number of other cases have been reported. The reported cases that we have been able to identify to date are summarized in Table 1 (6, 10–20).

The mean age at presentation for the reported patients including the present case is 33 years, with a range from 20 to 58 years. The majority presented in the second and third decades of life. No sex predominance is clear. For 5 (50%) of the 10 patients for whom such information was available, MFD was discovered during the workup of some unrelated trauma. Forty percent (including the present patient) presented with pain referable to the affected side. One patient presented with a compression fracture of the affected vertebra (12). Distribution of the lesions showed a slight preponderance of cervical (47%) over lumbar (40%) involvement.

To our knowledge, the present patient represents only the second report of pathology-proved MFD af-

TABLE 1. Monostotic Fibrous Dysplasia of the Spine

First author	Date	Age (yr)	Sex	Location
Schlumberger (11)	1946	20	M	C4
Ledoux-Lebard (12)	1953	58	F	L1
Rosendahl-Jansen (13)	1956	35	F	C4
Harris (6)	1962	42	M	L4
Daniluk (14)	1979	28	F	L4
Resnik (10)	1984	27	F	L4
Rosenblum (15)	1987	20	M	T1
Kahn (16)	1988	23	M	L3
Troop (17)	1988	NA	NA	Lumbar
Wright (18)	1988	NA	NA	C2
		NA	NA	C5
Hu (19)	1990	41	M	C2
Ehara (20)	1992	NA	NA	C1
		NA	NA	L3
Penrod (present study)	1993	40	F	T3
Total no. of cases with information available	15	10	10	15
		Range, 20–58 Mean, 33	5/10 M 5/10 F	7/15 Cervical 6/15 Lumbar 2/15 Thoracic

NA, not available.

fecting the thoracic region of the spine. The rarity of this type of lesion can make for a difficult diagnostic dilemma, making biopsy or surgical excision or both necessary. Mistaken diagnosis of osteoblastoma, osteoid osteoma, nonossifying fibroma, Paget's disease, aneurysmal bone cyst, and vertebral hemangioma has been reported (10, 18, 21).

MFD is typically managed conservatively once diagnosed. In the recently reported vertebral cases, however, excision of the lesion and some variety of stabilizing procedure were common (10, 15, 16, 18). Clearly, more aggressive management is indicated when the stability of the vertebral column is potentially compromised. Atlantoaxial dislocation recently was reported in a patient with polyostotic fibrous dysplasia involving C2 (21).

CONCLUSION

A 40-year-old patient had back pain aggravated by motion for 9 months. Imaging features were of a lytic well-circumscribed lesion of the right pedicle of T3. The present case report and the literature review serve to highlight the rarity of vertebral involvement with MFD and this report represent only the second such reported lesion of the thoracic region of the spine. Features of MFD in general and the diagnostic and treatment dilemmas unique to vertebral involvement have been discussed.

PATHOLOGICAL DIAGNOSIS

MFD of the pedicle at T3.

The authors wish to thank Eleanor Murphy for assistance with manuscript preparation.

REFERENCES

1. Feldman F. Tuberous sclerosis, neurofibromatosis and fibrous dysplasia. In Resnik D, Niwayama G (eds): Diagnosis of Bone and Joint Disorders. Philadelphia: WB Saunders, 1988;4057–4072.
2. Kransdorf MJ, Moser RP Jr, Gilkey FW. Fibrous dysplasia. Radiographics 1990;10:519–537.
3. Edeiken J (ed): Roentgen Diagnosis of Diseases of Bone. Baltimore: Williams & Wilkins, 1982;994–1027.
4. Schwartz DT, Alpert M. The malignant transformation of fibrous dysplasia. Am J Med Sci 1964;247:1–20.
5. Mirra JM, Piero P, Gold RH (eds): Bone Tumors. Philadelphia: WB Saunders, 1989;191–226.
6. Harris WH, Dudley R, Barry RJ. The natural history of fibrous dysplasia. J Bone Joint Surg [Am] 1962;44:207–233.
7. Dahlin D (ed): Bone Tumors. Springfield: Thomas, 1978;362–367.
8. Henry A. Monostotic fibrous dysplasia. J Bone Joint Surg [Br] 1969;51:300–306.
9. Firrat D, Stutzman L. Fibrous dysplasia of the bone: review of 24 cases. Am J Med 1968;44:421–429.
10. Resnik C, Lininger J. Monostotic fibrous dysplasia of the cervical spine: case report. Radiology 1984;151:49–50.
11. Schlumberger HG. Fibrous dysplasia of single bones (monostotic fibrous dysplasia). Milit Surg 1946;99:504–527.

12. Ledoux-Lebard G, Soulquin C. Les localisations vertébrales de la dysplasie fibreus des os ou maladie de Jaffé-Lichtenstein. Presse Med 1953;61:272–273.

13. Rosendahl-Jensen S. Fibrous dysplasia of the vertebral column. Acta Chir Scand 1956:6:490–494.

14. Daniluk A, Witwicki T. A case of fibrous dysplasia of the spine. Chir Narz Ruchu Ortop Pol 1979;44:165–167.

15. Rosenblum B, Overby C, Levine M. Handler M, Sprecter S. Monostotic fibrous dysplasia of the thoracic spine. Spine 1987;12:939–942.

16. Kahn A, Rosenberg PS. Monostotic fibrous dysplasia of the lumbar spine. Spine 1988;13:592–593.

17. Troop JK, Herring JA. Monostotic fibrous dysplasia of the lumbar spine: case report and review of the literature. J Pediatr Orthop 1988;8:599–601.

18. Wright JF, Stoker DJ. Fibrous dysplasia of the spine. Clin Radiol 1988;39:523–527.

19. Hu SS, Healy JH, Huvos AG. Fibrous dysplasia of the second cervical vertebra. A case report. J Bone Joint Surg [Am] 1990; 72:781–783.

20. Ehara S, Kattapuram SU, Rosenberg AE. Fibrous dysplasia of the spine. Spine 1992;17:977–979.

21. Nishiura I, Koyama T, Takayama S. Fibrous dysplasia of the cervical spine with atlanto-axial dislocation. Neurochirurgia 1992;35:123–126.